

University of Groningen

Main problems experienced by children with epidermolysis bullosa

van Scheppingen, Corinne; Lettinga, Ant T.; Duipmans, Jose C.; Maathuis, Care G. B.; Jonkman, Marcel F.

Published in:
Acta dermato-venereologica

DOI:
[10.2340/00015555-0376](https://doi.org/10.2340/00015555-0376)

IMPORTANT NOTE: You are advised to consult the publisher's version (publisher's PDF) if you wish to cite from it. Please check the document version below.

Document Version
Publisher's PDF, also known as Version of record

Publication date:
2008

[Link to publication in University of Groningen/UMCG research database](#)

Citation for published version (APA):

van Scheppingen, C., Lettinga, A. T., Duipmans, J. C., Maathuis, C. G. B., & Jonkman, M. F. (2008). Main problems experienced by children with epidermolysis bullosa: A qualitative study with semi-structured interviews. *Acta dermato-venereologica*, 88(2), 143-150. <https://doi.org/10.2340/00015555-0376>

Copyright

Other than for strictly personal use, it is not permitted to download or to forward/distribute the text or part of it without the consent of the author(s) and/or copyright holder(s), unless the work is under an open content license (like Creative Commons).

The publication may also be distributed here under the terms of Article 25fa of the Dutch Copyright Act, indicated by the "Taverne" license. More information can be found on the University of Groningen website: <https://www.rug.nl/library/open-access/self-archiving-pure/taverne-amendment>.

Take-down policy

If you believe that this document breaches copyright please contact us providing details, and we will remove access to the work immediately and investigate your claim.

Downloaded from the University of Groningen/UMCG research database (Pure): <http://www.rug.nl/research/portal>. For technical reasons the number of authors shown on this cover page is limited to 10 maximum.

CLINICAL REPORT

Main Problems Experienced by Children with Epidermolysis Bullosa: a Qualitative Study with Semi-structured Interviews

Corinne van SCHEPPINGEN^{1,2}, Ant T. LETTINGA², José C. DUIPMANS¹, Carel G.B. MAATHUIS² and Marcel F. JONKMAN¹

¹Center for Blistering Diseases, Department of Dermatology, and ²Center for Rehabilitation, University Medical Center Groningen, University of Groningen, Groningen, The Netherlands

The objective of this study was to identify and specify the problems of children with epidermolysis bullosa. The questions explored were: (i) What do children with epidermolysis bullosa experience as the most difficult problems; (ii) What is the impact of these problems on their daily life; and (iii) Do these experiences differ between mildly and severely affected children? Qualitative research methodology was used, comprising a series of semi-structured interviews with children with different (sub)types of epidermolysis bullosa. The interviews were analysed systematically with help of the qualitative software package Atlas-ti. Five main themes were found: (i) having an itchy skin, (ii) being in pain, (iii) having difficulties with participation, (iv) lack of understanding of others, and (v) the feeling of being different. Severely affected children suffered most from itch and treatment-related pain. Mildly affected children had more problems with activity-related pain. Mildly affected children also had more concerns about their appearance and the teasing and staring of others than did severely affected children. Both groups had difficulties with participation, the visibility of their disease and the feeling of being different. *Key words: chronic disease; children's problems; qualitative research.*

(Accepted August 27, 2007.)

Acta Derm Venereol 2008; 88: 143–150.

Marcel F. Jonkman, Center for Blistering Diseases, University Medical Center Groningen, Department of Dermatology, Hanzeplein-1, NL-9700 RB Groningen, The Netherlands. E-mail: m.f.jonkman@derm.umcg.nl

Epidermolysis bullosa (EB) is a rare skin disorder with a lifelong exceptional liability of the skin to blister after minor mechanical trauma. Due to a genetic abnormality, the different skin layers do not adhere adequately, so that the skin surface can break down easily after being hit, rubbed, scratched or during the process of swallowing. There are varying degrees of severity, ranging from mild forms with only local involvement of the skin surface and with no impact on overall longevity, to severe forms involving various organs with early postnatal death or chronic progression (1). It is estimated to affect about

one in 40,000 births (2). There are 3 principal groups of EB, based on the level at which the skin breaks down: EB simplex (EBS), junctional EB (JEB) and dystrophic EB (DEB). In EBS, blistering is often mild and lesions heal without scarring. JEB can present itself as widespread blistering of the skin and mouth at birth, which may be life-threatening. DEB varies from a mild dominant form with lesions on hands, knees and elbows, to a recessive form with generalized blistering and severe scarring that leads to mitten deformities of the hands and feet (1).

Children with EB can be confronted with extensive and complex problems, such as never-ending itch, repeatedly painful wound care, disfigurement, embarrassment and difficulties in meeting social demands (3–5). Nowadays, there appears to be increased attention focused on the physical disabilities and psychological implications that EB brings to children (3–6). Horn & Tidman (3), for example, investigated the quality of life of children with EB. Their results reflected the wide variety of problems encountered by EB sufferers. In this regard, it is quite a distressing result that the most severely recessive form of EB causes greater impairment of quality of life than any other skin disease previously assessed. Fine et al. (4) assessed the impact of different types and subtypes of EB on the activities of daily life, showing that children with all types of EB are likely to be at least partially dependent on others, especially in daily activities involving walking and personal hygiene. However, the majority of these studies tend to be quantitative in design, and little attention has been paid to the children's own beliefs about and experiences with their chronic illness (7–10). EB patients themselves occasionally write about growing up with EB and the problems they have experienced (5, 11). Sam Hall (5), a teenager with EB, for example, wrote about her problems with dressing changes, the accompanying pain and her unusual appearance. However, these stories are anecdotal.

This study is part of a two-sided research project in which the most difficult problems of both children with EB and their parents are explored (12). This part of the project has focused on the child's experiences. Thus, the aim of this study has been to gain a more qualitative and in-depth understanding of the wide-ranging and complex problems that children with EB have to deal

with. With this aim in mind 3 questions were included: (i) What problems do children with EB actually experience as being the most difficult? (ii) What is the impact of these problems on their daily life? and (iii) Are there differences in experiences between mildly and severely affected children? To address these questions we have adopted a qualitative research design in the form of semi-structured interviews with children with all types of EB.

MATERIALS AND METHODS

Participants

The children were recruited from the EB dermatology database of the University Medical Center Groningen, which consisted of 48 children from 6 to 18 years of age (September 2005). The criteria for eligibility included children who were diagnosed with a (sub)type of EB (13), who were aged between 6 and 18 years of age, who could speak and understand the Dutch language and who were willing and motivated to participate in the study (informed consent). The definitive sample comprised 9 families with 11 children with various types of EB. Five of the children were diagnosed with EBS type Köbner, Weber-Cockayne or Dowling-Meara (EBS-K, EBS-WC, EBS-DM), 2 with JEB non-Herlitz type (JEB-nH), 2 with dominant DEB (DDEB) and 2 with recessive DEB, Hallopeau-Siemens type (RDEB-HS). Three of their parents were also diagnosed with EB (Table I). Severe disease was defined as having generalized blistering with motion impairment and mild disease as having localized blistering or generalized blistering without motion impairment.

Procedure

An interview guide was developed through an extensive document analysis of EB, including literature into EB, patient's medical files, patient folders, and a booklet describing an extensive homecare program for EB patients. The interview guide was subsequently pilot-tested on 2 children with another chronic skin disease. Thereafter, some interview topics and leading questions were refined. The interview questions were open-ended and focused on children's daily activities, their hobbies, their school and social life, the treatment of their EB and their hopes for the future.

Prior to the acquisition of participants, the research was submitted to and approved by the local ethics committee. Thereafter, the researcher informed the children and their parents about the purpose and content of the study by means of an ex-

tensive letter. The researcher had a professional background in performing qualitative research and experience in interviewing on sensitive topics. Participation was voluntary and children and parents who agreed to participate were asked to sign a written consent form. The same researcher completed all interviews. The child interview took place in the home situation in order to reduce the impact of time-consuming travel, except for one case, which was located in the hospital.

After a period of "getting to know each other" an interview lasting from 60 to 100 minutes was conducted. Children were reassured that their personal stories would not be revealed to other people (14). The interviewer let the children know that they were the experts on their situation, clarifying that the interviewer did not know the answers already (14–16). During the interviews reflective listening was used to explore the meanings that children gave to specific words (17). Probes were constructed to suggest that other children might feel the same way and "action words" were used by asking what the child would do instead of how he or she felt in order to elicit the child's responses (18). Younger children were interviewed while playing a game, going for a walk or drawing, as most of these children's conversations also occur alongside other activities (7, 17). The interviewer took notes on the child's condition, the interviewer-child interaction, and the home environment to complement the spoken data. In addition, the interviewer visited some children in the hospital during their stays and documented the observations there.

At the end of the interview the children were given the opportunity to summarize the most difficult problems that they experienced. To that effect the children filled in a 5-point scale with clouds, ranking the various problems that they had mentioned in the interview from a bit difficult (sun behind the cloud) to most difficult (thundercloud) (Fig. 1). This 5-point scale was used to assess the impact of the various problems from the child's own point of view. The children who were able to read were sent a transcribed copy of the interview, which gave them a chance to reflect on the interview report.

Data analysis

The interviews were tape-recorded and transcribed verbatim afterwards. All data were analysed in their original language to preserve the original meanings. The transcribed interviews were subjected to content analysis with the qualitative software package Atlas-ti (19). Atlas-ti offers tools for managing, extracting, comparing, exploring and reassembling meaningful pieces of text from large amounts of data in a systematic way. With the help of this software package, quotations that were relevant with regard to the aim of the study were coded based on the meaning and judgment that the children ascribed to their problems. The

Table I. Background data for children with epidermolysis bullosa (EB)

Family no.	Age of child (years)	EB subtype	Severity	Number of siblings	Family characteristics	Affected parents
1	17	JEB-nH	Mild	1	Two parents	
2	11	RDEB-HS	Severe	1	Two parents	
3	14	DDEB	Mild	0	Separated parents	Mother (DDEB)
4	10	EBS-K	Mild	1	Separated parents	Father (EBS-K)
	12	EBS-K	Mild	1	Separated parents	Father (EBS-K)
5	12	EBS-WC	Mild	2	Separated parents	
6	9	EBS-DM	Severe	6	Two parents	Mother (EBS-DM)
	17	EBS-DM	Severe	6	Two parents	Mother (EBS-DM)
7	13	RDEB-HS	Severe	5	Two parents	
8	14	DDEB	Mild	2	Two parents	
9	6	JEB-nH	Mild	1	Two parents	

JEB-nH: junctional EB non-Herlitz type; RDEB-HS: recessive dystrophic EB, Hallopeau-Siemens type; DDEB: dominant dystrophic EB; EBS-K: EB simplex type Köbner; EBS-WC: EB simplex type Weber-Cockayne; EBS-DM: EB simplex type Dowling-Meara.

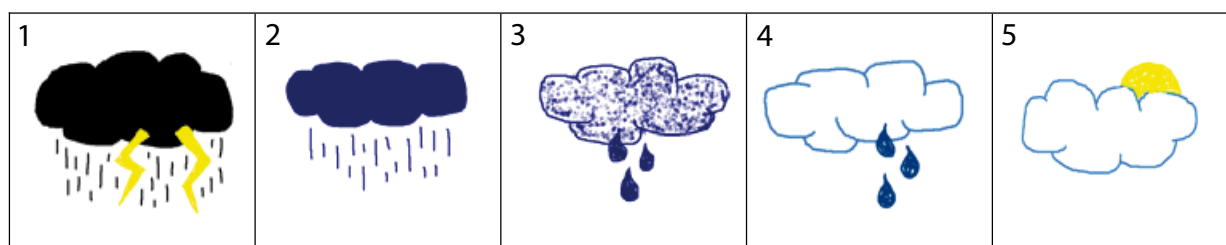


Fig. 1. Cloud ranking scale for children with epidermolysis bullosa.

interview records were read carefully and then coded into small categories, such as “problems with the visibility of the disease” and “not being able to follow fashion trends.” These categories were further categorized into main themes, in this context “concerns about appearance.” The interview fragments were continuously compared in order to identify new themes and to specify the complex data in clear categories. Finally, the categories and main themes were described to a second researcher, who was new to the results of the study. Indistinctness between categories was discussed by the 2 researchers, resulting in a refinement of the themes and categories.

The problems the children ordered on the cloud ranking scale were written down in their own vocabulary. In the subsequent analysis the researcher rephrased overlapping problems using summarizing themes. For example, when the child formulated problems such as “pain in the eye” and “pain on the chest” in the problem list, this was summarized as the problem of “being in pain.” One child did not complete the scale as she was too young to understand (6 years old). So, in the end 10 children completed the cloud ranking scale. Table II depicts how the children defined and ranked their main experienced problems on the scale.

RESULTS

Most of the children were motivated and took their time putting into words what it was like and how it felt to have EB. As a result, the children discussed various and wide-ranging problems in the interviews. The problems the children with EB described were ordered into 5 main themes: (i) having itchy skin, (ii) being in pain, (iii) having difficulties joining in with others, (iv) the visibility of the disease, and (v) the feeling of being different. Within these 5 themes, differences in experience were found between mildly and more severely affected children (Table II).

Having itchy skin

All children with severe (sub)types of EB put the problem of itching at the top of the cloud ranking scale. These children mentioned that they really suffered from itchy skin. One severely affected child stated: “*I think that the itch is so awful that you can’t even begin to understand. (...) I wish I could just stop it, but I can’t.*” The children mentioned 2 causes for their itchy skin. The first cause was the skin itself: dry skin and healing wounds contributed to their feelings of itch. The weather was also frequently mentioned as a contributing factor. A warm, sunny, sweaty day with high humidity was experienced as very unpleasant.

In the interviews, the question was raised as to what the itch *did* to the children so that it was experienced as such a problem. First of all, it appeared that the itch was demanding because of the vicious circle that it could induce. That is, the itch usually provoked a need to scratch, which could irritate the skin even more, worsen the existing wounds or create new blisters. This then could aggravate the feelings of itch all the more. Moreover, it was especially the *continuous duration* of the itch that was experienced as a great burden. A severely affected child stated: “*It’s almost always there, wherever I am.*” The observations during the interview confirmed this statement, as the children were restlessly moving in their chairs most of the time, trying to scratch their backs or rubbing over their dressings.

Not only was the itch physically troublesome, the children also experienced it as a continuous psychological burden, as they mentioned that they constantly had to keep themselves from scratching. The itch could pro-

Table II. Five main problems of children with different types of epidermolysis bullosa (EB), ranked on the cloud scale

	Problem of the child (n=10)	Subtype EB (ranking 1–5)
1	Having itchy skin	EBS-DM (1), RDEB-HS (1), RDEB-HS (1)
2	Being in pain – Fear of pain	JEB-nH (1), DDEB (4), RDEB-HS (2), RDEB-HS (2) DDEB (1)
3	Having difficulties joining in with others	EBS-DM (2), EBS-WC (1), EBS-K (1), DDEB (2), DDEB (3), RDEB-HS (4), RDEB-HS (3)
4	Visibility of the disease – Being teased/ stared at – Concerns about appearance	EBS-K (2), DDEB (1), DDEB (2) EBS-K (2), EBS-K (1), JEB-nH (2), DDEB (1)
5	Feelings of being different – Lack of understanding by others	EBS-DM (3) EBS-DM (4), DDEB (3)

1 = most difficult problem (thundercloud), etc. (see Fig. 1). For abbreviations see Table I.

voke a constant struggle within the child not to scratch. When this did not succeed, the scratching behaviour could give them a feeling of low self-control. One child: *"I try as hard as I can (not to scratch) because I really know that scratching isn't good. (...) But sometimes it just happens, that's what's so awful."*

Furthermore, the itch also had social consequences for the children. They mentioned, for example, that the itch kept them from having a good night's sleep: *"Even in your sleep you're all tensed up, telling yourself, 'Don't scratch!'"* As a result, they could be very tired during the day, which disrupted their planned daily activities. For some children the itch also led to concentration problems at school: *"I just couldn't follow it (the lesson) anymore, just itch, itch, all the time."*

Not only were the children affected by the skin itching, but the parents were also involved. One of the severely affected children mentioned that he frequently asked his parents to get up at night to scratch his itchy skin when he could not sleep. Furthermore, it annoyed some children that their parents "policed" them all day long. That is, the parents were continuously watching the child and warning about scratching behaviour.

In contrast, the mildly affected children did not put itching on their EB problem list. They mentioned that they could itch sometimes, when for example their wounds were healing. But it was not of continuous duration, nor did it put any extreme demand on their self-control or on the relationship with their parents.

Being in pain

Four children put the problem of pain on the cloud ranking scale. The children consequently described 2 different types of pain, which can best be characterized as *treatment-related* pain and *activity-related* pain. To begin with, the most severely affected children would describe one type of pain, which seemed to be the hardest for them, the treatment-related pain resulting from their dressing changes. These children were covered from head to toe with bandages, which had to be changed almost every day by their parents and care providers. This ritual could take more than 2.5 h; getting the (sticky) bandages off, bathing, cleaning the wounds, removing new blisters and dressing the whole body again. One severely affected child stated: *"Because the dressings stick to the wounds and they have to come off. That hurts a lot."* Another child, despite being very open about his experiences in the interviews, refused to discuss his dressing changes, saying: *"That's something I really don't want to talk about. The dressing changes are something I really don't like."*

In this context, observations in the home and the hospital brought about a related problem, namely the children's fear of pain during treatment and medical procedures.

Some children started screaming and crying before parents or healthcare professionals even started their treatment. Other children fearfully asked questions in order to find out exactly what was going to happen or they tried to keep a tight control by giving strict orders to the parent or physician about what he/ she should or should not do.

The severely affected children also described a kind of *activity-related* pain, that is, pain that could interfere with certain activities, such as eating a nice meal or going for a walk. Some mildly affected children also mentioned this kind of *activity-related* pain. One of them stated: *"When you're walking in the city, you just aren't comfortable walking. I can't imagine walking without pain."* One of them also talked about her fear of pain, not caused by treatment of the blisters, but as a result of being too active, which made her very careful. The child stated: *"Sometimes the fear of certain kinds of pain takes over. (...) But that's because in your mind you expect it to be worse than it is."* She even ranked her fear of pain higher than the pain itself.

Although both mildly and severely affected children experienced this activity-related pain, it seemed that the pain sensations for mildly affected children were less intense than for severely affected children. Some mildly affected children even stressed that they could get used to the pain, for example, pain coming from already existing wounds. One of them stated: *"I just see it, like, the pain is just there and that's all. I hardly feel it. (...) Actually, it's just normal."* In this context, some of these children mentioned that they could endure their pain easily because they perceived a higher pain threshold than others did. One child stated: *"When I tear my whole leg open people react like they're going to faint. And then I just keep on walking. (...) You get used to it, I think."* In contrast with the mildly affected children, the severely affected children could not get used to their pain during repetitive wound care. For them this was an *everyday* and *unavoidable* pain.

Difficulties in joining in with others

The children with the most severe EB mentioned that they sometimes had a strong wish to perform activities which they knew were not possible for them anymore because of their physical limitations. One of them: *"I would just like to run free for once. I can't do that anymore because of my feet."* The limitations of the hands due to progressive fusion of the fingers also caused frustrations. One boy stated: *"(I don't like) when I drop things all the time and then I can't pick them up."* For another severely affected child the fact that she couldn't eat anymore was difficult, especially when there was a strong smell of nice food in the house: *"Sometimes it smells so good, like a hot dinner at home ... then you want some too and that just doesn't work."*

On the other hand, those children with the somewhat milder types of EB did not have these kinds of physical limitations, so they were able to participate in more activities than the severely affected children could. The most troubling thing for them was not the physical limitations themselves, but the fact that they could not join in with certain enjoyable activities, such as gymnastics or soccer, because of the risk of developing blisters, being hit or falling down. One of them gave quite a moving description when she was asked how it felt to be unable to join her friends: *"(I feel) sort of left out."* Being unable to join in gave her the feeling of being an outsider.

Although the children with the mildest types of EB were able to participate in almost all activities, they stated that it was their own problem that they always had to be careful when doing things. One of them stated: *"In fact you do get involved in stuff, but you just have to be careful all the time."* He even found "having to be careful" the most difficult aspect of his EB. He did not like the fact that he first had to take precautions with his bandages when going skating, for example, before he could join his friends.

Moreover, some children talked about having to make difficult choices; having to weigh the enjoyable activities with friends against the possible consequences: blisters and pain. One of them described this as the most demanding problem with her EB; that is, she had to keep her illness in mind constantly. In addition, making the decision to join in or not join in with certain activities was hindered by a feeling of the *uncontrollability* of the disease, as one of them said: *"You really can't say, 'Today I'm going to have problems,' because you don't know that, you just always have to wait and see."* At the same time, other children emphasized that the consequences of just doing things they liked, but if they were smart, they probably should not be doing, did not bother them that much. When they made the decision to participate, then they *"caused"* the blisters themselves. It seemed that when these children had a sense of control over their EB, then the consequences of being incautious (the wounds and blisters) were less frustrating than when the blisters and wounds appeared spontaneously.

Visibility of the disease

Most of the children seemed to be bothered by the reactions of those in their immediate surroundings to the "visibility" of their EB. One of them stated: *"I find it kind of hard that when I hurt myself you can see I have this disease."* That is, most of the mildly affected children had visible blisters or scars on their hands, faces or other body parts. Despite the fact that the skin of children with severe types of EB was almost entirely covered in clean bandages, their body was obviously

affected even more. They could have large scabs, scar tissue or open and smelly wounds all over their heads, while their hands and feet could be disfigured.

The "visibility" of the children's skin disorder brought about 2 main problems that were quite frequently put on the cloud ranking scale, for example, the staring and teasing of other people. Some mildly affected children seemed to experience people staring as one of the most frustrating aspects of their EB. One of them stated: *"The most annoying thing is when people look at you and they don't say a word. Then you know that they're just staring all the time."* But also severely affected children disliked people looking at them: *"I just get crazy with all that staring."* Most of the children used critical responses to this staring such as, *"Is there something wrong," "Can I help you" or "Just go on playing."* Nevertheless, one of the mildly affected children emphasized that it was more in their own minds that others were always looking at them: *"Actually they don't pay that much attention to it, but (you think they do)."* In short, on the one hand the problem was the blatant staring by strangers, while on the other hand their greater awareness or sensitiveness was a contributing factor in their feeling of being stared at.

Those children with the milder types of EB also told a lot of stories about other people trying to hit them on purpose or calling them names like "wart-man". One boy mentioned that nobody wanted to touch him until he explained that his disease was not contagious: *"If I don't explain it, they run away."* As a result of this discrimination some children withdrew from activities where they knew they ran the risk of being teased, such as swimming or taking a shower with soccer mates. It became clear that the teasing and unwanted questions were difficult for the children to cope with.

The way most children dealt with their concerns about the visibility of their disease was by trying to hide their wounds. One young boy, for example, always tried to be the first in line at the swimming pool, so nobody could see the wounds on his stomach. Or, otherwise, he used his hands to hide the wounds. For him this was the most difficult thing about his skin disease: *"That I have to hide it."* Another girl recognized that she automatically put her affected hands in her pockets or inside the long sleeves she was wearing on purpose. Although hiding the disease could give the children some relief, it could also make the illness "invisible." One mildly affected child mentioned that it was just this "invisibility" that made others lack any understanding of the impact of EB on her life: *"They don't see it, so they never understand what I actually have."*

In contrast to the teasing the mildly affected children experienced, the children with the most severe types of EB mentioned that they did not get teased that much. Although they did get unpleasant remarks sometimes, it seemed that these did not bother them too much. One

of them: *"Sometimes they call me a 'mummy'. But then I say 'Yeah, fun isn't it.'"* Also strangers staring, which they also found quite irritating, did not stop them from continuing their activities. One of the children stated: *"It doesn't bother me that much I don't go outside anymore. It's not that bad."* They also did not discuss problems related to their appearance, such as shame or fear of the visibility of their disease. However, from the interview observations it became clear that they really tried to look nice by wearing attractive clothing over their dressings and glamorous shoes and by using fluorescent bandages. It could be that in this way they were also trying to distract attention from their unusual and messy appearance.

Feelings of "being different"

One mildly affected boy said on the telephone that there was no point talking about the problems he experienced with his skin disease because he was just a "normal" boy like all the others his age. However, in the interview with his mother she mentioned various problems that he encountered with his EB. He could not go to school independently or use the keys to open the door. One of the interviewed children described her life with EB in a similar, but less extreme manner: *"I try to do everything the others do as much as I can. So, there are only a few little things where I'm different from an average school kid."* But also for her, living with EB entailed a lot of limitations, as she later described.

Because the rare skin disorder, EB was rather unknown to most of the people in their immediate surroundings, some children experienced a lack of understanding regarding their illness, which could make them feel alone in the experience they faced. In this context, children did not like having to explain their EB over and over. One of them: *"Sometimes I find it hard that I always have to explain it to everybody (...) Because you have this idea, 'I'm just normal,' and 'I just want to be somebody without a skin disease.'"*

Although they wanted to be seen as "normal," most of the children felt that they were somehow different. One of them stated: *"That's what I wish for, just to be a normal kid, because I'm kind of different now."* Moreover, it seemed that it also bothered some children that others identified them as different. One severely affected child stated: *"When people, like, have to describe me, they say 'that girl with the skin disease.' (...) I would rather have people see me as myself instead of 'that girl with the skin disease.'"* In the end, the wish to be (seen as) normal appeared to be closely related to all the problems discussed above. Depending on the individual child, it entailed the wish to be without pain and itch; to be able to join in activities with "normal" children; to not be teased, stared at and have to explain EB all the time; to be without any unusual appearance, or to have an identity unrelated to the skin disease.

DISCUSSION

Recent literature focusing on the problems of children with EB has already given some idea of the impact and extent of their difficulties (3–5). For example, Horn & Tidman (3) investigated their quality of life. According to their study, severely affected children had major difficulties with sports, with symptoms such as pain and itch and with the treatment of the skin. Children with the milder types of EB showed the highest scores on questions about symptoms, being embarrassed, self-conscious or sad, and difficulties with going out, playing or hobbies.

Although these studies give us more insight into the main difficulties of children with different types of EB, they leave many questions unanswered. For example, it remains unclear *why* these difficulties are ranked so high or, more specifically, *which* aspects are the most annoying. It has also been left unclear what the *impact* of these difficulties is on the everyday lives of children with EB. Although our study has to be seen as explorative in design, the small sample size made it possible to deliver an in-depth understanding of the problems of children with EB. From our study, for example, it became clear that it was the unbearable itch that was the most frustrating for children with severe types of EB and that this entailed a physical, a psychological, as well as a social burden in their everyday lives. Mildly affected children, however, did not experience their itch as the main problem since it was not continuously present.

Fine et al. (4) found, as regards the pain experience of children with various types of EB, that they all mostly suffered from cutaneous pain when walking or standing. Some more severely affected children were also in pain during other daily activities. However, the authors did not put their pain in a broader context. Our study has identified that the most severe pain for severely affected children was not related to daily activities, but to their treatment. That is, the severely affected children suffered from intense pain during their dressing changes, which was often even too difficult to discuss. They could not get used to this everyday and unavoidable pain. On the other hand, children with the milder types of EB were more bothered by activity-related pain, where it interfered with doing things they liked to do. In addition, both mildly and severely affected children discussed their fear of pain. It seemed as if the fear of pain and the accompanying stress resulted in exacerbating the pain experience. In contrast to the severely affected children, some mildly affected children reported that they could get used to their pain and that they experienced a higher pain threshold than others.

Regarding the previously described major problems of children with EB as far as sports and going out, playing and hobbies (3) are concerned, our study has specified what children found most difficult about these things. We discovered that, depending on the severity

of the EB, the most difficult thing in the context of sports and playing was the physical limitations (severe EB), being unable to participate with friends (milder EB) and constantly having to be careful (mildest EB). In this context, Tackett et al. (20) stated that the major problems of children with all kinds of physical disabilities were social rather than physical. In our study, the problems of the mildly affected children indeed confirm this statement. However, the severely affected children did find the physical limitations themselves, such as being unable to walk or eat, to be their major problem. It seemed that for them the physical disabilities were so burdensome that the social consequences were of minor importance.

Rumsey et al. (21) already found that visible disfigurement could lead to major problems concerning contact with others: social interactions, exposing the disfigurement to other's gaze and displays of ignorance and negative comments by others. In the context of EB, Horn & Tidman (3) discovered that *both* mildly and severely affected children had the lowest scores on questions about teasing, bullying, being avoided or being asked questions and about contact with friends. This, in part, is in contradiction with our findings.

Although children with the milder types of EB were quite invisibly affected, we discovered that they did suffer from being teased and picked on. Moreover, they seemed to have more problems with that than did children with the most severe types of EB. The more severely affected children couldn't hide their illness that easily, since they were always covered with dressings or sitting in a wheelchair. It might be that as a result they had learned to deal with the staring and the remarks. That is, these children are so visibly different that they might have been forced to "accept" themselves as a person with a disability (22).

Other studies so far confirm that people with extensive disfiguration are not likely to have more (appearance) problems than those with only minor differences (21). Furthermore, the contradiction with the study of Horn & Tidman (3) might be a result of the fact that the questionnaire took only the problems of the previous week into consideration. In our interviews, children described that some situations in the distant past still had an impact on their present behaviour. For example, they mentioned not going swimming any longer because of previous unwanted questions, or not taking a shower with friends because of their staring. So, existing problems might have been overlooked when only the present week was considered, since they appear to have their origin in the history of the child's illness.

Several studies show that children with all kinds of chronic illnesses can suffer from a feeling of being different (7, 23–27). Literature on the problems of children with EB does not describe the feeling of being different as a major burden for these children (3–6). However,

we found that the physical pain of their condition, the lack of understanding by others in relation to what they were going through, the inability to join in with others and their unusual appearance could all cause a feeling of being different from others. Regarding the relative rarity of the EB skin disorder, this feeling might be even stronger for children with EB than for children with other chronic illnesses. For this reason, trying to participate, to camouflage, and to explain EB, e.g. "it's not contagious", in order not to differ from others are rather important aspects for these children when dealing with their illness. Taken together, the present study revealed that the main problems of children with EB are itch, pain and disfigurement, which is perceived as contagious, resulting in constrained social acceptance. This combination of problems is unique to EB, differentiating it from other chronic illnesses.

ACKNOWLEDGEMENTS

This study was funded by shared grants from the Johanna Kinderfonds, the BIO Kinderrevalidatie Fund and the Kinderrevalidatie Fonds Adriaanstichting (The Netherlands). We gratefully acknowledge the participation of the children with EB in our study, their willingness to respond to the many questions and the time they took to put into words their often difficult experiences.

Conflicts of interest: None to declare.

REFERENCES

1. Fine JD, Bauer EA, McGuire J Moshell A, editors. Epidermolysis bullosa. Clinical, epidemiologic, and laboratory advances, and the findings of the national epidermolysis bullosa registry. Baltimore: Johns Hopkins University Press, 1999.
2. Horn HM, Priestley GC, Eady RAJ, Tidman MJ. The prevalence of epidermolysis bullosa in Scotland. *Br J Dermatol* 1997; 136: 560–564.
3. Horn HM, Tidman J. Quality of life in epidermolysis bullosa. *Clin Exp Dermatol* 2002; 27: 707–710.
4. Fine JD, Johnson LB, Weiner M, Suchindran C. Assessment of mobility, activities and pain in different subtypes of epidermolysis bullosa. *Clin Exp Dermatol* 2003; 29: 122–127.
5. Hall S. Life, epidermolysis bullosa and chasing tornadoes. *J Wound Care* 2004; 13: 405–406.
6. Andreoli E, Mozetta A, Angelo C, Paradisi M, Foglio Bonda PG. Epidermolysis bullosa. psychological and psychosocial aspects. *Dermatol Psychosom* 2002; 3: 77–81.
7. Alderson P, Sutcliffe K, Curtis K. Children as partners with adults in their medical care. *Arch Dis Child* 2006; 91: 300–303.
8. Papadoulos L, Bor R, Legg C. Psychological factors in cutaneous disease: an overview of research. *Psychol Health Med* 1999; 4: 107–126.
9. Eiser C, editor. Growing up with a chronic disease. The impact on children and their families. London: Jessica Kingsley Publishers, 1993.
10. La Greca AM. Social consequences of pediatric conditions: fertile area for future investigation and intervention? *J Ped Psychol* 1990; 15: 285–307.

11. Dietz, MJ. A day in the life of a patient with DDEB. *J Am Acad Dermatol* 2004; 51: S58–S59.
12. Scheppingen, C, Lettinga AT, Duipmans JC, Maathuis CGB, Jonkman MF. Main problems of parents of a child with epidermolysis bullosa. *Qual Health Res* (in press).
13. Fine JD, Eady RAJ, Bauer EA, Briggaman RA, Bruckner-Tuderman, L, Christiano, A, et al. Revised classification system for inherited epidermolysis bullosa: report of the second international consensus meeting on diagnosis and classification of epidermolysis bullosa. *J Am Acad Dermatol* 2000; 42: 1051–1066.
14. Docherty S, Sandelowski, M. Focus on qualitative methods: interviewing children. *Res Nurs Health* 1999; 22: 177–185.
15. Ross MR, Ross SA. The importance of type of question, psychological climate and subject set in interviewing children about pain. *Pain* 1984; 19: 71–79.
16. Garbarino J, Stott FM, Faculty of the Erikson Institute. Guidelines for interviewing children. In: Garbarino J, Stott FM, Faculty of the Erikson Institute, editors. *What Children Can Tell Us: Eliciting, Interpreting, and Evaluating Critical Information from Children*. San Francisco, Calif.: Jossey Bass, 1992. p. 170–202. ISBN: 978-1-55542-465-7.
17. Bricher G. Children and qualitative research methods: a review of the literature related to interview and interpretive processes. *Nurs Res* 1999; 6: 65–77.
18. Faux SA, Walsh M, Deatrick J. Intensive interviewing with children and adolescents. *W J Nurs Res* 1988; 10: 180–194.
19. Muhr T, Friese S, editors. *Atlas-ti. The knowledge workbench. Visual qualitative data analysis and knowledge management in education, business, administration and research*. USA Bohemia, Long Island: Research Talk Inc., 2004.
20. Tackett P, Kerr N, Helmstadter G. Stresses as perceived by children with physical disabilities and their mothers. *J Rehab* 1990; 56: 30–34.
21. Rumsey N, Clarke A, White P, Wyn-Williams M, Garlick W. Altered body image: appearance-related concerns of people with visible disfigurement. *J Adv Nurs* 2004; 48: 443–453.
22. Thompson RJ, Gustafson KE, editors. *Adaptation to chronic childhood illness*. Washington: American Psychological Association, 1990.
23. Enskär K, Carlsson M, Golsäter M, Hamrin E, Kreuger A. Life situation and problems as reported by children with cancer and their parents. *J Ped Oncol Nurs* 1997; 14: 18–26.
24. Woodgate RL. Adolescent's perspectives of chronic illness: "It's hard." *J Ped Nurs* 1998; 13: 210–223.
25. D'Auria JP, Christian BJ, Richardson LF. Through the looking glass: children's perceptions of growing up with cystic fibrosis. *Can J Nurs Res* 1997; 29: 99–112.
26. Velsor-Friedrich B, Vlasses F, Moberly J, Coover L. Talking with teens about asthma management. *J School Nurs* 2004; 20: 140–148.
27. Tracey JP. Growing up with chronic illness: the experience of growing up with cystic fibrosis. *Hol Nurs Pract* 1997; 12: 27–35.